D-TRANSPOSITION OF THE GREAT ARTERIES

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DEFINITION TGA: **AORTA** ARISES FROM THE **RIGHT** VENTRICLE
“CORRECTED” OR “L” TRANSPOSITION

- Ao arises from RV
- Ventricles are switched
- RV is systemic ventricle; LV pulmonary ventricle
- RV failure over time
- Tricuspid valve can be abnormal, hence TR
- Heart block
Discordant ventricular-arterial relationship

Aorta arising from Right Ventricle and Pulmonary artery from Left Ventricle

Usual form = D-TGA, aorta sits anterior and rightward and originates from RV

20-30 per 100,000 live births

Strong male predominance (60-70%)

Extracardiac and genetic malformations uncommon (<10%) compared to other congenital heart disease
ASSOCIATED CV ABNORMALITIES

- Atrial septal defect
- Ventricular septal defect (40%)
- LV Outflow tract obstruction, PS (10-15%)
- RV outflow tract obstruction, AS/CoA (10%)
- TGA in complex hearts:
  - Tricuspid Atresia
  - Single left ventricle (DILV)
  - L-TGA (congenitally corrected TGA)
NEED FOR PRENATAL DIAGNOSIS IN TGA

- TGA is a form of CHD where there are dramatic changes after birth & separation from placenta

- Potential for hemodynamic compromise is great, and immediate BAS and PGE can be life saving

- Controlled environment for delivery, transfer to skilled, experienced team for cardiorespiratory management offers the best potential outcome
FLOW PATTERNS: IN THE FETUS

RV

LV

Streaming Via Ductus Venosus

Ao

PA

LUNGS

BODY

DUCTUS ART
CONSEQUENCES OF ALTERED FETAL FLOW PATTERNS IN TGA

- Lower O2 content blood to the brain
- Higher O2 content to the DA
- Relatively decreased blood flow volume across DA (arises from LV instead of RV)
- DA smaller than normal or restrictive
- Smaller DA may reduce blood volume through LV, decrease atrial level shunt
- Atrial level restriction?
FLOW PATTERNS AFTER BIRTH:
OPEN INTER-ATRIAL COMMUNICATION

- RV
- LV
- PA
- LUNGS
- Ao
- BODY
- PDA
FLOW PATTERNS: PDA & RESTRICTIVE ATRIAL SEPTUM

RV → PA → LUNGS

Ao → BODY

PDA
NORMAL GREAT ARTERY RELATIONSHIP: CROSSING VESSELS
CROSSING VESSELS?
PARALLEL VESSELS
PARALLEL VESSELS
LONG AXIS – GREAT VESSELS

- RV
- AO
- PA
- LV
LONG AXIS – GREAT VESSELS
TGA, VSD, PS
TGA, VSD, PS
TGA, VSD, PS
ONCE TGA DETECTED, OTHER KEY FEATURES...

- Ductus Arteriosus
- Foramen Ovale
DUCTUS ARTERIOSUS
DUCTUS ARTERIOSUS
WIDE OPEN INTER-ATRIAL COMMUNICATION
SMALL PFO
BALLOON ATRIAL SEPTOSTOMY
(RASHKIND PROCEDURE)
How best to plan for delivery?
“Fetuses with D-TGA should be delivered in a hospital that can manage the hypoxia and hemodynamic compromise that can occur...”
**CHOP Delivery Classification Scheme for CHD**

<table>
<thead>
<tr>
<th>CLASS</th>
<th>Definition</th>
<th>Examples</th>
<th>Action</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>no hemodynamic instability</td>
<td>VSD, AV Canal, “pink” TOF</td>
<td>evaluation</td>
</tr>
<tr>
<td>II</td>
<td>moderate severity, including ductal dependant lesions, stable hemodynamics</td>
<td>Pulmonary atresia, Critical Coarct or AS, HLHS</td>
<td>PGE</td>
</tr>
<tr>
<td>III</td>
<td>possibility or likelihood of hemodynamic instability.</td>
<td>TGA</td>
<td>PGE + Cardiac ICU involvement</td>
</tr>
<tr>
<td>IMPACT</td>
<td>hemodynamic instability is anticipated at separation from placental circulation. [<em>Immediate post-partum access to cardiac therapy (IMPACT)</em>]</td>
<td>HLHS + IAS, Ebstein’s, CHB, hydropic fetus</td>
<td>C-section in Cardiac facility</td>
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Transposition of the Great Arteries (TGA) 
Arterial Switch Operation - Step 1

AO = Aorta
PA = Pulmonary Artery
LA = Left Atrium
RA = Right Atrium
LV = Left Ventricle
RV = Right Ventricle

Left and Right Coronary Arteries Removed From AO on “buttons”
PDA Divided
Aorta and PA Divided
Transposition of the Great Arteries (TGA)
Arterial Switch Operation - Step 2

- Pulmonary Artery Moved in Front of Aorta
- Aorta Reconstructed
- Coronary Arteries Connected to Aorta on "Buttons"
- Right Coronary Artery

AO = Aorta
PA = Pulmonary Artery
LA = Left Atrium
LA = Left Atrium
RA = Right Atrium
LV = Left Ventricle
RV = Right Ventricle

- Oxygen-rich Blood
- Mixed Blood
- Oxygen-poor Blood
- Mixed Blood
There are a number of coronary artery patterns in D-TGA.

Infants with an intramural course, i.e. an artery traveling within the wall of the vessel (G, H), are at increased risk for mortality after the arterial switch procedure.

From: Echocardiography in Pediatric and Congenital Heart Disease: From Fetus to Adult, edited by Lai, Mertens, Cohen, and Geva
TGA REPAIR – ARTERIAL SWITCH OPERATION
Arterial Switch Operation

• Excellent Outcomes
• > 98% survival
• Long term concerns
  • Coronary insufficiency?
  • Aortic root dilation
Cardiovascular Outcomes After the Arterial Switch Operation for D-Transposition of the Great Arteries

Paul Khairy, MD, PhD; Mathieu Clair, MD; Susan M. Fernandes, MHP, PA-C; Elizabeth D. Blume, MD; Andrew J. Powell, MD; Jane W. Newburger, MD, MPH; Michael J. Landzberg, MD; John E. Mayer Jr, MD

- 400 pts arterial switch between 1983-1999
- 26 (6.5%) perioperative deaths
- In perioperative survivors, overall and arrhythmia free survival was

![Probability of arrhythmia or sudden death](image)
TRANSPOSITION OF THE GREAT ARTERIES

- A form of congenital heart disease in which dramatic improvement in outcome has taken place over the past 30 years.
- It is expected that most infants born today with TGA will survive into adulthood.
- Coronary artery insufficiency and aortic root dilation are potential complications that will require follow-up.